Cytological analysis of ascitic fluid from an adolescent male with intra-abdominal desmoplastic small round cell tumor

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**Background**: Desmoplastic small cell tumor (DSRCT) is a rare malignancy mainly involving the peritoneum of adolescent males. Only a few reports on the cytology of body fluid from patients with DSRCT have been documented.

**Case**: We present the cytological findings for an ascitic fluid sample taken from a 13-year-old Japanese male with intra-abdominal DSRCT. The patient was admitted to our hospital for abdominal distention and gross ascites. A cytological evaluation of the ascitic fluid revealed the presence of monotonous small round cells and a small number of another kind of large atypical cells with thick cytoplasm corresponding to the characteristic perinuclear round hyaline material of DSRCT. The tumor was resected, but the patient died from a recurrence two years later.

**Conclusion**: DSRCT should be considered as a differential diagnosis of gross ascites accompanied by a bulky intra-abdominal tumor in adolescents.

**Key words**: Desmoplastic small round cell tumor—Cytology—Ascites

I. Introduction

Desmoplastic small round cell tumor (DSRCT) was first identified as a unique morphological feature of small round cell malignancies in two cases reported independently in 1989. DSRCT arises most frequently from the peritoneum and omentum of adolescent males. The clinical course is characterized by a high recurrence rate and rapid progression, usually with a lethal outcome⁴. We previously reported a case of intra-abdominal DSRCT with gross ascites but did not describe the cytological findings for the ascitic fluid⁵. Cytological examination of the ascitic fluid is an important first step in the diagnosis of intra-abdominal tumors. Although at least 80 cases of DSRCT have been reported to date, only a few authors have described cytological findings for body fluids⁶. We therefore present the cytological findings for an ascitic fluid sample taken from our patient.
II. Case Report

A 13-year-old Japanese boy presented with general fatigue and nausea in September 1989. Ascites and a bulky intra-abdominal tumor were observed upon admission in September 1989. Continuous drainage of the ascitic fluid was instituted; the maximal volume was 2 L/day. Following the resection of the tumor, intensive chemotherapy with Adriamycin, cyclophosphamide and cisplatinum was administered. The pathological diagnosis was intraabdominal DSRCT. Two years later, the patient again developed abdominal distension. A relapse of DSRCT with ascites was diagnosed, and he died in October 1991. An autopsy was not permitted.

III. Pathological Findings for the Ascitic Fluid Sample

Cytological Findings

A Papanicolaou-stained smear specimen of ascitic fluid exhibited a large number of small atypical cells that were up to two times larger than the peripheral lymphocytes. These cells exhibited a scanty cytoplasm and round to oval nuclei with irregular contours, hyperchromasia and prominent nucleoli. Small clusters of these atypical cells, resembling those of small cell carcinoma in the lung, were often observed. In addition, a small number of another kind of atypical cell with a large amount of thick cytoplasm that stained light green was observed (Photo. 1a). The cytoplasm of these cells was negative for PAS staining. A Giemsa stain did not suggest hematopoietic cell features in either the small or the large atypical cells. The remaining cellular components of the ascitic fluid were collected and embedded in paraffin. Sections stained with hematoxylin and eosin (H & E) exhibited numerous undifferentiated round cells with perinuclear round hyaline structures in the cytoplasm that resembled the larger atypical cells in the smear specimen. These cells corresponded to the tumor cells observed in the resected tumor tissue (Photo. 1b, 3).

Immunohistochemical Findings

An immunohistochemistry examination of paraffin sections from cell blocks was performed using the labelled streptavidin-biotin complex (LSABC) method with an anti-keratin, wide-spectrum antibody (DAKO, Kyoto, Japan), an anti-swine vimentin antibody (DAKO), an anti-neuron-specific enolase (NSE) antibody (DAKO), and an anti-desmin antibody (DAKO). Around 30% of the tumor cells in the ascitic fluid were positive for vimentin, NSE and desmin (Photo. 2), but negative for wide keratin in the cytoplasm. The sub-cellular distributions of these proteins were not clear, including that of the cytoplasmic globular structure.

Histological Findings

During the operation, the tumor seemed to originate from the omentum. A tumor specimen weighing 4980 g was resected. The tumor exhibited a gyrus-like surface and was a grayish white color. The cut surface was white, solid and fibrous. Microscopically, the tumor was comprised of tumor-cell nests and broad-banded stroma (Photo. 3a). A large number of tumor cells were round or polygonal and epithelioid, with bean-shaped or oval nuclei in eccentric locations and a relatively abundant cytoplasm containing pale perinuclear round material (Photo. 3b) The electron microscopic structure of this material corresponded to that of the perinuclear whorls of cytoplasmic intermediate filaments. Several smaller tumor cells with atypical round nuclei and scanty cytoplasm were also observed.

IV. Discussion

Since tumor cells with cytoplasmic globular inclusions were not predominant in the ascites specimens, the cytological finding of small round atypical cells first suggested neuroblastoma, embryonal rhabdomyosarcoma, Ewing's sarcoma, primitive neuroectodermal tumor (PNET), malignant lymphoma and small cell carcinoma as differential diagnoses of this tumor. The PAS-negative
Photo. 1  a: Smear specimen prepared from ascitic fluid. Monotonous small round cells with irregularly shaped and hyperchromatic nuclei are visible. A few cells with large amounts of thick cytoplasm that stains light green can be seen (Papanicolaou staining, ×40).  b: Cell block examination of the ascites. Poorly differentiated small round cells with perinuclear round hyaline material are visible (H&E staining, ×40).

Photo. 2  Immunostaining for vimentin (a), NSE (b) and desmin (c). The tumor cells are strongly positive (×40).

cytoplasm suggested that a myogenic tumor and Ewing's sarcoma were unlikely. In addition, the existence of a small number of another kind of atypical cell with a large amount of thick cytoplasm that stained light green implied a polymorphism that would probably exclude Ewing's sarcoma, PNET and small cell carcinoma. The Giemsa stain findings were useful in ruling out malignant lymphoma. However, further investigation was difficult because the immunohistochemistry on the smeared slides showed a high background. Thus, we adopted the cell block method. Sections stained with H&E exhibited numerous undifferentiated round cells with a perinuclear round hyaline structure in the cytoplasm. The tumor cells in the cell blocks were occasionally positive for vimentin, NSE and desmin in the cytoplasm, and the frequencies of
immunoreactivity were less than those of the resected specimen. However, the sub-cellular distribution of these proteins, including the cytoplasmic globular structure, was not clear (Photo. 2). At this time, we narrowed the differential diagnoses to malignant rhabdoid tumor and DSRCT. During the histological examination of the resected specimen, paranuclear whorl structures consisting of intermediate filaments and dot-like regions of desmin and vimentin positivity were revealed by electron microscopy. These findings distinguished the tumor from other small round cell tumors, and we finally diagnosed the tumor as DSRCT after considering the clinical features, which included a bulky intra-abdominal tumor in a male adolescent with no other tumors found in the visceral organs, unlike the typical findings for malignant rhabdoid tumor.

In conclusion, the diagnosis of DSRCT using body fluid cytology might be difficult if the larger atypical cells with the characteristic cytoplasm of this tumor are a minor component of the cytological features of an ascitic fluid specimen dominated by small round atypical cells. Therefore, the consideration of DSRCT as a differential diagnosis for bulky intra-abdominal tumors in adolescents with gross ascites should not be overlooked. Recently developed molecular methods as well as immunohistochemistry techniques, including antibodies for desmin, may be helpful in the diagnosis of DSRCT using ascites fluid samples.

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